A131.E1223
JACC March 9, 2010
Volume 55, issue 10A

QUALITY OF CARE AND OUTCOMES ASSESSMENT

PROGNOSTIC VALUE OF A SYSTEMATIC FAMILIAL SCREENING IN IDIOPATHIC DILATED CARDIOMYOPATHY. THE EXPERIENCE OF TRIESTE HEART MUSCLE DISEASE

ACC Poster Contributions
Georgia World Congress Center, Hall B5
Sunday, March 14, 2010, 9:30 a.m.-10:30 a.m.

Session Title: Genomics and Myocardial Phenotypes
Abstract Category: Genetics and Clinical Outcomes
Presentation Number: 1029-166

Authors: *Marco Merlo, Michele Moretti, Giulia Barbati, Dario Gregori, Andrea Di Lenarda, Luisa Mestroni, Gianfranco Sinagra, Ospedali Riuniti and University, Trieste, Italy*

Background: Familial screening of patients with dilated cardiomyopathy (DCM) allows an early diagnosis of the disease in family members. It is unclear if familial forms of DCM (FDC) have a different long-term outcome with respect to the sporadic DCM. The aim of this study was to compare long-term prognosis of FDC with respect to sporadic forms in order to assess the role of familial screening.

Methods: From 1988 to 2007 we enrolled 637 consecutive DCM patients in the Trieste Heart Muscle Disease Registry. We compared non-proband FDC patients (NP-FDC) with a sample of 96 sporadic forms, randomly matched by year of enrolment (2:1) in order to increase the efficiency of comparisons.

Results: FDC were 130 (20.4%), 82 (12.9%) were probands. The familial screening effectively diagnosed DCM in 48 (7.5%) NP-FDC patients. With respect to the random sample, NP-FDC patients at enrollment were younger (40±16 vs 47±13 years, p=0.002), less severely symptomatic (NYHA class III-IV 8% vs 28%, p=0.006), had higher left ventricular ejection fraction (35±10 vs 30±9%, p=0.005). The survival free from heart transplant at 2, 5 and 10 years was respectively 93, 91 and 82% in NP-FDC patients versus 86, 76 and 62% of sporadic forms (p=0.04). After stratification for NYHA classes, no difference in survival has been observed between sporadic and NP-FDC patients.

Conclusions: FDC represented 20% of our population. Familial screening of DCM patients allowed early recognition of 7.5% of patients. NP-FDC patients which had less advanced disease at presentation and a better long-term outcome.